abcam

Product datasheet

Anti-UROS antibody ab58097

1 Image

Overview

Product name	Anti-UROS antibody
Description	Mouse monoclonal to UROS
Host species	Mouse
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human
Immunogen	Recombinant full length protein, corresponding to amino acids 1-266 of Human UROS

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: None PBS, pH 7.2
Purity	Protein G purified
Clonality	Monoclonal
Isotype	IgG1
Light chain type	kappa

Applications

Our Abpromise guarantee covers the use of **ab58097** in the following tested applications.

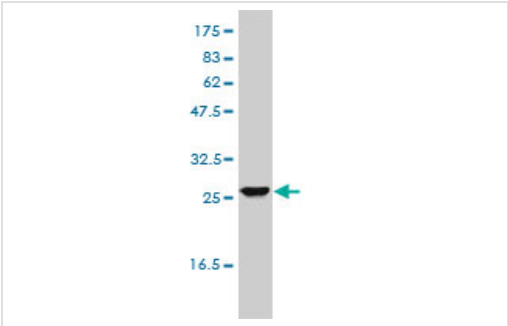
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		Use a concentration of 1 - 5 µg/ml. Predicted molecular weight: 29 kDa.

Target

Function	Catalyzes cyclization of the linear tetrapyrrole, hydroxymethylbilane, to the macrocyclic uroporphyrinogen III, the branch point for the various sub-pathways leading to the wide diversity of porphyrins. Porphyrins act as cofactors for a multitude of enzymes that perform a variety of processes within the cell such as methionine synthesis (vitamin B12) or oxygen transport (heme).
Tissue specificity	Ubiquitous.
Pathway	Porphyrin metabolism; protoporphyrin-IX biosynthesis; coproporphyrinogen-III from 5-aminolevulinate: step 3/4.
Involvement in disease	Defects in UROS are the cause of congenital erythropoietic porphyria (CEP) [MIM:263700]; also known as Gunther disease. Porphyrias are inherited defects in the biosynthesis of heme, resulting in the accumulation and increased excretion of porphyrins or porphyrin precursors. They are classified as erythropoietic or hepatic, depending on whether the enzyme deficiency occurs in red blood cells or in the liver. The manifestations of CEP are heterogeneous, ranging from nonimmune hydrops fetalis due to severe hemolytic anemia in utero to milder, later onset forms, which have only skin lesions due to cutaneous photosensitivity in adult life. The deficiency in UROS activity results in the non-enzymatic conversion of hydroxymethylbilane (HMB) into the uroporphyrinogen-I isomer. Note=Severe congenital erythropoietic porphyria is associated with non-immune hydrops fetalis, a generalized edema of the fetus with fluid accumulation in the body cavities due to non-immune causes. Non-immune hydrops fetalis is not a diagnosis in itself but a symptom, a feature of many genetic disorders, and the end-stage of a wide variety of disorders.
Sequence similarities	Belongs to the uroporphyrinogen-III synthase family.

Images



Western blot - Anti-UROS antibody (ab58097)

UROS antibody (ab58097) at 1ug/lane + HL-60 cell lysate at 25ug/lane.

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit https://www.abcam.com/abpromise or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors